Congenital Insensitivity to Pain and Anhidrosis (CIPA) and Anesthesia

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Hereditary sensory and autonomic neuropathy type IV, also known as congenital insensitivity to pain with anhidrosis CIPA, is a rare hereditary syndrome. The syndrome is one of the inherited disorders grouped under Hereditary Sensory and Autonomic Neuropathies (HSAN). The syndrome affects the ectodermal structures such as skin, nervous system and bone. The presentation of the syndrome is unique as it can cause marked sweat gland dysfunction, episodic hyperpyrexia secondary to high environmental temperature, insensitivity to pain, self-inflicted injury and intellectual disability. The anesthetic management of the CIPA is challenging, which could lead to preoperative complications.

We present a case of rare hereditary sensory and autonomic neuropathy syndrome type IV related to anesthesia. An eighteen-year-old female, diagnosed as a case of congenital insensitivity to pain and anhidrosis syndrome was admitted electively for external fixation of a fracture of the right elbow. The patient was diagnosed on genetic testing at 10 months and confirmed the presence of mutations in neurotrophic tyrosine kinase receptor type 1 (NTRK1) gene.

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